JACC Vol. 17, No. 2
February 1991:48A

Monday, March 4, 1991
4:00PM-5:30PM, Room 260, West Concourse
Progress in Congenital Heart Disease Diagnosis

4:00

ECHOCARDIOGRAPHIC EVALUATION OF ATRIOVENTRICULAR VALVE ORIFICE ANATOMY IN CHILDREN WITH ATRIOVENTRICULAR SEPTAL DEFECT

L. LuAnn Minich, A. Rebecca Snider, Roger P. Vermilion, Edward L. Bove, University of Michigan, Ann Arbor, Michigan

In atrioventricular septal defect (AVSD), the common atrioventricular valve can have a common orifice or can be divided by bridging leaflet tissue into two separate orifices. To determine the accuracy of a two-dimensional echocardiographic technique that we devised for evaluation of the number of atrioventricular valve orifices, we prospectively examined all children (n=69) undergoing surgical repair of AVSD from 4/87 to 8/90. The presence of bridging leaflet tissue and the number of atrioventricular valve orifices were determined using a subcostal imaging plane. From a standard subcostal four-chamber view, the plane of sound was rotated 30 to 45° clockwise until the atrioventricular valve was seen *en face*. The plane of sound was then tilted from a superior to an inferior direction so that cross-sectional views of the atrioventricular valve were examined from the inferior margin of the atrial septum to the superior margin of the ventricular septum. Of the 69 patients, 6 patients (9%) were excluded due to failure to obtain the appropriate subcostal images (3 due to obesity and 3 due to operator failure). The remaining 63 children ranging in age from 1 day to 13.5 years and in weight from 1 to 55 kg. constituted the study group. Echocardiographic results were compared to surgical observations in 62 patients and to autopsy findings in 1 patient. With the two-dimensional echocardiographic technique, 32 of 33 patients with a common atrioventricular orifice and 28 of 30 patients with 2 separate atrioventricular orifices were correctly identified. By chi-square analysis, the echocardiographic techniques allowed correct identification of a common orifice with 94% sensitivity and 97% specificity. For correct identification of two separate orifices, the echocardiographic technique had 97% sensitivity and 94% specificity. The positive predictive values of the echocardiographic technique were 97% for a common orifice and 93% for two separate orifices. Thus, in patients with AVSD, the presence of bridging leaflet tissue and the number of atrioventricular valve orifices can be accurately determined using a subcostal two-dimensional imaging plane.

4:15

INTRAVASCULAR ULTRASOUND IMAGING FOR EVALUATING THE ANATOMY OF RIGHT VENTRICULAR OUTFLOW TRACT AND PULMONARY ARTERY LESIONS AS A GUIDE FOR INTERVENTIONAL CATHETERIZATION STUDIES IN PEDIATRIC PATIENTS. David J. Sahn, Francois Ricou, Abraham Rothman, Richard Swensson, Kyung Chung. Univ of Calif, San Diego, CA.

We used a new 4.8 French 20MHz ultrasound catheter (Diasonics-IVUS) to image 17 pts (ranging in wt from 5.2-56 Kg) to evaluate the capability of intravascular ultrasound (IVUS) for imaging the structure of RV outflow, pulmonary valve, and PA lesions as a guide for interventional catheter treatment. The scanning catheter was easily introduced through 6FR venous sheaths or used over-the-wire through 7FR sheaths. The structure and severity of PA constrictions was localized in 3 pts with post-shunt PA obstructions (pulmonary atresia or tetralogy), 1 pt with congenital multifocal PA constrictions, 2 pts with PA bands, and 1 with a conduit obstruction; all with close correlation to angiographic diameter). The distortion of PA wall structure induced by scar at the site of stenosis, the multi-layered nature of conduit "peel", and the imaging of subvalvar dynamic obstruction was achieved. The internal mural anatomy of (muscular) subclavian artery and Gortex shunts and their sites of narrowing could be defined as could the muscular wall structure of PDA. Three pts were imaged before and after balloon valvuloplasty for pulmonary stenosis with documentation of the change in PV orifice size corresponding to their hemodynamic relief. A complex Fontan pt was documented by IVUS to have had a left pulmonary artery embolus with organized thrombotic material visualized in the left PA tree. IVUS catheters are now suitable for use in young children and can provide anatomical insights which may be helpful in guiding catheter interventional therapy for congenital heart disease pts.

4:30

ANATOMIC TYPES OF LEFT VENTRICULAR OUTFLOW TRACT OBSTRUCTION IN COMMON ATRIO-VENTRICULAR CANAL

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Narrowing of the left ventricular outflow tract (LVOT) is a known anatomic feature of common atrio-ventricular canal (CAVC). Since the type of LVOT obstruction will affect the surgical management of pts with CAVC, we sought to determine the anatomic types and their relative frequency. We analyzed 96 postmortem heart specimens with CAVC, 68 complete CAVC, 28 incomplete CAVC and 70 normal controls. LVOT obstruction was present in 43/96 (45%): 27/68 (40%) with complete CAVC and 16/28 (57%) with incomplete CAVC. There were 3 anatomic types of LVOT obstruction: attachment of atrioventricular valve (AVV) leaflet to septum in 16/27 complete CAVC and 15/16 incomplete, extension of anterolateral papillary muscle into LVOT in 1/16 incomplete, and malalignment of the posterior portion of the infundibular septum toward the left in 11/27 complete CAVC - isolated (1), associated with AVV attachment (8), with papillary muscle extension into LVOT (1), and with both papillary extension and AVV attachment (1).

While typical attachment of AVV leaflet to ventricular septum was the predominant cause of LVOT obstruction in CAVC in general, leftward deviation of the posterior aspect of the infundibular septum into the LVOT contributed to obstruction in 41% of complete CAVC cases with LVOT obstruction. Therefore the absence of typical AVV attachment to ventricular septum seen in CAVC does not rule out the presence of LVOT obstruction. Infundibular septal malalignment and abnormal papillary muscle extension into LVOT are also significant causes of subaortic stenosis in CAVC.

4:45

UNRECOGNIZED SEGMENTAL PULMONARY STENOSES AND LOBAR PULMONARY VASCULAR DISEASE IN TETRALOGY OF FALLOT (TOF) Nancy D Bridges, Stanton B Perry, John F Keane, Valerie Mandell, James E Lock. The Children's Hospital, Boston, MA.

Partial isolation of portions of the pulmonary bed, a feature of TOF (especially with pulmonary atresia (PA)), may cause vascular disease in the unobstructed lung. Five patients (pts) (4 TOF/PA, 1 TOF) had reduced flow to a large part of the lung due to severe lobar &/or segmental pulmonary artery stenoses. Flow was predominantly to the right middle lobe (RML) & left lung in 1 pt; the RML and right lower lobe in 1 pt; the right upper lobe in 1 pt, and the RML in 2 pts. Outflow tract reconstruction (with VSD closure in 2) resulted in overperfusion and elevated mean pressures (33, 28, 37, 32, & 25 mm Hg) in the nonstenosed arteries. Progressive symptomatology & cyanosis due to vascular disease in perfused regions occurred in all; in each, the potential for recruitment of pulmonary vessels and stabilization of vascular disease went unrecognized. All pts had transcatheter dilation of multiple segments; 3 pulmonary artery stents were placed in one pt. Pressure past the stenoses increased from 17 +/- 11 mm Hg to 33 +/- 19 mm Hg (p=0.02) after dilation. Subjective exercise tolerance improved in all; ventricular ectopy resolved in 1; one previously inoperable pt has had complete repair.

Pulmonary hypertension in TOF or TOF/PA should prompt an exhaustive search for segmental and lobar stenoses. Stenosed areas are recruitable, & failure to do so may result in progressive vascular disease in overperfused segments.